

CASE REPORT

Castleman's Disease of the Neck

Chih-Cheng Chen¹, Rong-San Jiang¹, Guan Chou², Ching-Ping Wang^{1*}

*Departments of ¹Otolaryngology-Head and Neck Surgery and ²Pathology,
Taichung Veterans General Hospital, Taichung, Taiwan, R.O.C.*

Castleman's disease is an uncommon benign disease of unknown etiology that induces reactive lymph node hyperplasia. There are 2 histologic subtypes of Castleman's disease: hyaline vascular type and plasma cell type. Tissue biopsy is necessary for a definitive diagnosis. Castleman's disease of the neck occurs rarely. On reviewing the literature, most Castleman's disease of the neck presented with a single mass. We report here an 80-year-old man with 2 neck masses of 1 year's duration. A 4-cm firm movable mass in the left submandibular area and another 2-cm firm movable mass in the left level IV area were noted. The patient was diagnosed with Castleman's disease of the neck after resection of 1 of the masses. There was no evidence of progression or recurrence of the disease after 21 months of follow-up. We suggest that this was a rare case of multicentric cervical Castleman's disease. [*J Chin Med Assoc* 2007;70(12):556-558]

Key Words: Castleman's disease, lymph node hyperplasia, neck mass

Introduction

Castleman's disease was first described by Benjamin Castleman,¹ who presented 13 cases of localized mediastinal lymph node hyperplasia resembling thymoma. It is a rare benign disease of unknown cause, giving rise to lymph node hyperplasia. Noninvasive techniques are usually inadequate for precise diagnosis. Accurate diagnosis depends on histopathologic evaluation after surgical resection. The neck is an uncommon site for Castleman's disease. From a review of the literature, most cases of Castleman's disease of the neck presented with a single mass. We report an 80-year-old man with 2 neck masses of 1 year's duration. He was diagnosed with Castleman's disease of the neck after resection of 1 of the masses. The clinical presentation, management and pathological characteristics of this disease are discussed, together with a literature review.

Case Report

In June 2004, an 80-year-old man with a left submandibular mass for 1 year presented to our clinic. A firm, movable and non-tender mass measuring 4 cm in diameter in the left submandibular area and another

firm, movable mass measuring 2 cm in diameter in the left level IV area were found. Ear, nose, nasopharynx, oropharynx, larynx, and other systemic examinations yielded normal findings. Fine needle aspiration of the 2 masses revealed benign lesions. Computed tomography (CT) of the neck showed a 4 × 3-cm homogeneous well-defined mass (Figure 1A) located in the left submandibular area and another 2 × 1-cm mass in the left level IV area (Figure 1B). The results of complete blood count, serum biochemistry and chest X-ray were normal. Under the impression of suspected lymphoma, we recommended the patient to receive excisional biopsy for the level IV mass. Under local anesthesia, the neck was explored via a cervical incision. The mass was found lying between the sternocleidomastoid muscle and left internal jugular vein. It was well encapsulated and firm. The mass was easily removed with minimal blood loss.

Macroscopically, it appeared to be a well-circumscribed and firm ovoid mass. The resection specimen consisted of 1 lymph node measuring 3.5 × 2 cm in diameter. Microscopically, it showed vascular proliferation within lymph node hyperplasia. Immunostaining including L26, OPD and bcl2 stains were all negative and the diagnosis of lymphoma was excluded. The pathologic findings were compatible with Castleman's

*Correspondence to: Dr Ching-Ping Wang, Department of Otolaryngology, Taichung Veterans General Hospital, 160, Section 3, Chung-Kang Road, Taichung 407, Taiwan, R.O.C.
E-mail: entcpw@gmail.com • Received: March 21, 2007 • Accepted: September 11, 2007

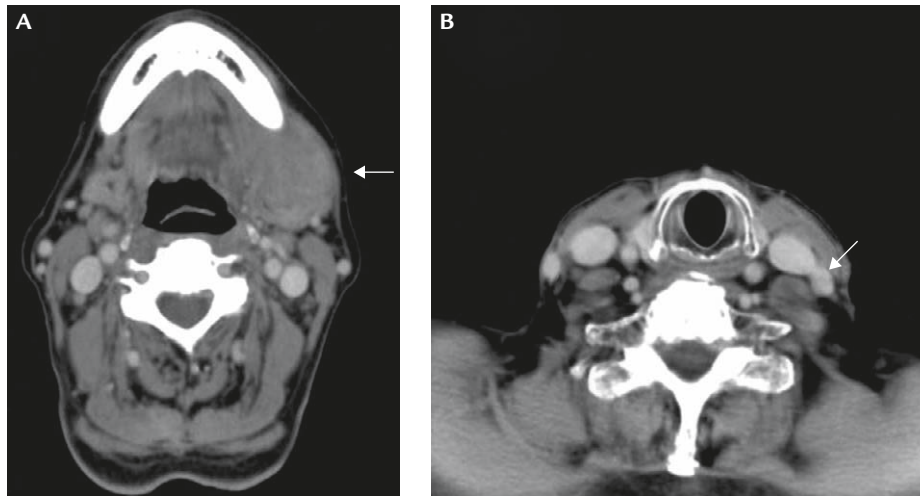


Figure 1. Computed tomography shows: (A) enlargement of the left submandibular gland, measuring about 4 cm in diameter; (B) lymph node of the left level IV neck area measuring about 2 cm in diameter and enhancement with contrast (arrow).

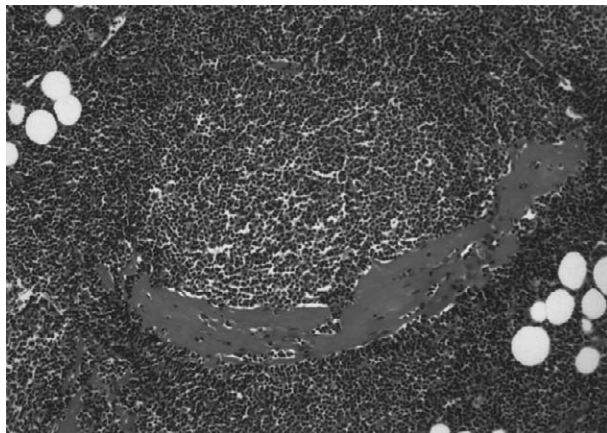


Figure 2. Hyaline vascular type: follicle. Capillaries with fibrous hyalinization penetrate the small follicle center. Note the capillary proliferation in the nearby interfollicular tissue (hematoxylin & eosin, 40 \times).

disease of hyaline vascular type (Figure 2). We suggested another resection for the left submandibular mass, but the patient did not consent due to his old age. There has been no evidence of progression of the left submandibular mass or recurrence after 21 months of follow-up.

Discussion

Castleman's disease is a benign lymphoproliferative disorder that was first described by Castleman and Towne in 1954.¹ It is characterized by enlarged hyperplastic lymph nodes usually presenting as a localized mass. Castleman's disease may occur at any age, but most occur in adults. It is also known as angiofollicular

lymph node hyperplasia, giant lymph node hyperplasia, angiofollicular hamartoma, and benign lymphoma.^{2,3} There are 2 histologic subgroups of Castleman's disease: hyaline vascular (80–90% of cases) and plasma cellular type. The hyaline vascular type is characterized by small lymphoreticular follicles scattered in hyper-vascular hyalinized stroma. The plasma cellular type is rare but more aggressive, characterized by mature plasma cell clusters between lymph follicles. The pathological findings in our patient were compatible with the hyaline vascular type. The most prominent sites include thorax (63%), abdominal cavity (12%) and axilla (4%)³ without systemic symptoms. Rarely, it can occur in the stomach.⁴ Casper suggested an algorithm for the evaluation and management of patients suspected to have Castleman's disease in 2005.⁵ Excisional biopsy is still an essential procedure for diagnosis. CT is another useful tool for ruling out Castleman's disease from lymphoma or thymoma.⁶ The lesions of Castleman's disease demonstrate enhancement on CT scan, but lymphoma and thymoma do not. In our case, both of the neck masses showed enhancement under contrasted CT scan. One of the masses was proven to be Castleman's disease, which is compatible with Patel et al's opinions⁶ (Figure 1B).

The neck is an uncommon site for Castleman's disease. Reviewing the literature, we found only a few cases of cervical Castleman's disease, and most of them presented with a solitary neck mass only.^{7,8} Multicentric cervical Castleman's disease is rare; only Osma et al⁹ have previously described a patient with 2 lymph nodes removed from the neck with Castleman's disease. In our case, the level IV mass was shown to be Castleman's disease and the submandibular mass remained the same

size after 21 months of follow-up. There was no evidence of progression or recurrence of the disease. Both masses showed enhancement under contrast-enhanced CT. We suggest that this patient was another rare case of multicentric cervical Castleman's disease. However, further excisional biopsy is essential to be sure of this diagnosis.

There is neither age prominence nor a sex predilection for the disease. Surgery is the treatment of choice in cases of giant lymph node hyperplasia, whereas chemotherapy, radiotherapy, and steroids are proposed for the multiple forms. No recurrences have been reported in the literature after complete resection of the hyaline vascular type. Sanz et al¹⁰ reported the only case of the plasma cell type in the head and neck, which recurred after 11 months. Our case showed no progression or recurrence over 21 months of follow-up.

References

1. Castleman B, Iverson L, Menerdez VP. Localized mediastinal lymph node hyperplasia resembling thymoma. *Cancer* 1956;9: 822–30.
2. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. *Cancer* 1972;29:670–83.
3. Altinli E, Pekmezci S, Balkan T, Somay A, Buyukbese MA, Tasci H, Akcal T. Castleman's disease masquerading as sigmoid colon tumor and Hodgkin lymphoma. *Swiss Surg* 2002;8: 7–10.
4. Chou SP, Chen LK, Lo SS, Li AF, Chang FC, Hwang S. Hyaline-vascular variant of Castleman's disease mimicking a gastric submucosal tumor. *J Chin Med Assoc* 2004;67:152–5.
5. Casper C. The aetiology and management of Castleman's disease at 50 years: translating pathophysiology to patient care. *Br J Haematol* 2005;129:3–17.
6. Patel U, Forte V, Taylor G, Sirkin W. Castleman's disease as a rare cause of a neck mass in a child. *J Otolaryngol* 1998;27: 171–3.
7. Somdas MA, Ketenci I, Bicer S, Senturk M, Guney E. Castleman's disease as an unusual neck mass: case report. *Ann Otol Rhinol Laryngol* 2004;113:459–61.
8. Beraldo S, Altavilla G, Bernante P, Pelizzo MR. Castleman's disease as an uncommon cause of a neck mass. *Acta Otolaryngol* 2006;126:108–11.
9. Osma U, Cureoglu S, Yaldiz M, Topcu I. Castleman's disease (giant lymph node hyperplasia) of the neck: a case report. *Eur Arch Otorhinolaryngol* 2001;258:42–4.
10. Sanz C, Sierra J, Cobarro J, Avellaneda R, Montserrat E, Rozman C. An unusual case of Castleman's disease restricted to the neck. *ORL J Otorhinolaryngol Relat Spec* 1992;54: 331–3.